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Title: A painful vegetating finger

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Clinical Findings

A 55-year-old female presented with a six-week history of inflammation, pain and paraesthesiae involving the left ring finger. The fingernail had been removed due to an assumed unresponsive bacterial infection 25 days prior to presentation. Her medical history included pemphigus vulgaris (PV), diabetes mellitus and β -thalassemia trait.

There was circumferential inflammation, desquamation and a purulent exudate with erythema at the leading edge suggesting an advancing lesion (Fig. 1). Intraorally, there was extensive ulceration on the dorsum tongue and a single ulcer involving the right buccal mucosa. All other skin and mucosal surfaces were unaffected.

Comprehensive investigations were undertaken. Bacterial skin swab revealed a heavy growth of *Citrobacter Koseri*. Viral swab was negative. Cortical irregularity of proximal phalanx was noted on a radiograph but osteomyelitis was subsequently excluded on MRI. Serum ELISA studies DsG1 of 6U/ml (negative) and DsG3 of 120U/ml (strongly positive).

Histopathological findings

An incisional biopsy showed fragments of necrotic debris with collections of neutrophils and numerous bacterial colonies. Clusters of acantholytic cells were also seen. There was no viable epidermis present on evaluation. There was no evidence of psoriasis in the tissue section examined. Gram stain highlighted both Gram-positive and gram-negative cocci. PAS, Grocott, Ziehl Neelsen and Wade Fite stain were negative (Fig. 2).

What is your diagnosis?

Diagnosis:

Pemphigus vegetans involving the digit

Discussion:

The patient was admitted for debridement and antiviral and antibacterial therapy pending culture results. She was treated with daily potassium permanganate soaks, topical corticosteroids (clobetasol 17-propionate 0.05%), oral azathioprine 150mg and prednisolone 10mg (Fig. 3). There was complete resolution within eight weeks (Fig. 4).

Pemphigus vegetans is an uncommon clinical subtype of PV thought to affect 1-5% of all pemphigus patients¹. Patients present with vegetating plaques composed of excessive granulation tissue and crusting which may evolve into exudative, vegetating masses with pustules and fissures. The intertriginous areas, scalp, and face are the most common sites affected. Oral involvement is common².

Classically, two subtypes of pemphigus vegetans are recognised³. Pemphigus vegetans of Neumann is characterised initially by typical PV lesions with vesicles and bullae that rupture to form hypertrophic granulating erosions, evolving into vegetating plaques. Pemphigus vegetans of Hallopeau is a milder form characterised by pustular lesions that, after rupturing, merge and evolve into vegetating erosions and are not preceded by bullae. The aetiology of pemphigus vegetans is unclear but it has been linked to drugs including captopril and heroin. Patients with known PV can transform into the vegetans subtype⁴.

Histologic examination shows hyperkeratosis, papillomatosis and prominent acanthosis.

Immunofluorescence findings in pemphigus vegetans are indistinguishable from those of PV.

DIF demonstrates deposition of IgG and C3 on the cell surface of keratinocytes. Indirect

immunofluorescence reveals circulating intercellular IgG in the majority of cases. In this case,

there was an increase in anti-DG3 titres from 104U/ml to 120U/ml that coincided with her presentation.

Citrobacter Koseri is an uncommon Gram-negative aerobic intestinal bacillus that does not usually reside in skin⁵. It is known to infect chronic ulcers and in this case may have secondarily infected the finger.

The course for pemphigus vegetans is similar to that of PV. Systemic glucocorticoids are the treatment of choice with addition of immunosuppressive agents to allow a steroid-sparing effect. Topical antiseptics such as potassium permanganate soaks and sodium hypochlorite 2% soaks have been shown to reduce granulation tissue formation⁶. Topical corticosteroids are commonly used for cutaneous lesions.

In conclusion, a focal vegetative lesion may present a diagnostic challenge. Pemphigus vegetans should always be included in the differential diagnosis in those with a background of PV. This is a rare case of pemphigus vegetans involving a digit.

Learning points

- PV can develop into pemphigus vegetans. Clinicians should be aware of this when a patient presents with a vegetative lesion.
- Biopsy is necessary to exclude other conditions that may present similarly such as acrodermatitis of Hallopeau (a form of psoriasis).

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CPD Questions

Learning objective

To understand the clinical and immunopathological features of pemphigus vegetans and how it relates to PV.

Question 1

The diagnosis of pemphigus vegetans may mimic which of the following conditions?

- (a) Pyodermatitis–pyostomatitis vegetans
- (b) Acrodermatitis of Hallopeau
- (c) Bacterial infection
- (d) Fungal infection
- (e) All of the above

Question 2

Which of the following are recognised treatments of pemphigus vegetans?

- (a) Anti-viral therapy, topical corticosteroids
- (b) Anti-bacterial measures, topical corticosteroids
- (c) Anti-bacterial measures, topical corticosteroids, systemic immunosuppression
- (d) Systemic immunosuppression
- (e) Topical corticosteroids

Answers

Question 1: (e)

Question 2: (c)

Figure Legends

Figure 1: Circumferential inflammation and desquamation with erythema at the leading edge

Figure 2: Fragments of necrotic debris with clusters of acantholytic cells and collections of neutrophils (A); Acantholytic cells, admixed with neutrophilic debris and cocci colonies (B) (H&E x60); Numerous colonies of Gram negative cocci (C) (Gram stain, x 60).

Figure 3: Day seven post debridement and application of clobetasol 17-propionate 0.05%

Figure 4: Eight weeks post therapy